Sclerosing Stromal Tumor of the Ovary

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Sclerosing stromal tumors are benign ovarian neoplasms of the sex cord stromal category which occur predominantly in the second and third decades of life. Herein, we report a 26-year-old woman who developed a sclerosing stromal tumor of the ovary with irregular menses but normal hormonal status. She was suspected to have a malignant tumor and underwent bilateral oophorectomy. Other ovarian stromal tumors include fibroma and thecoma which tend to occur in the fifth and sixth decades of life. It is, therefore, necessary to keep in mind the possibility of a sclerosing stromal tumor in a young woman.

Keywords: Sclerosing stromal tumor • sex cord stromal tumor of the ovary

Introduction

Sclerosing stromal tumors (SSTs) are rare benign ovarian neoplasms of the sex cord stromal category. Most cases occur predominantly in the second and third decades of life, and they are unilateral and well-circumscribed. Most patients present with nonspecific symptoms related to an adnexal mass. The tumor, with rare exceptions, is hormonally inactive. If it is hormonally active, it is usually androgenic and most frequently occurs during pregnancy. All reported cases of SST have been benign. Diagnosis of SST is often made by postoperative pathologic examination. The important differential diagnoses are other sex cord stromal tumors including fibroma, thecoma, and lipid cell tumors.

Case Report

A 26-year-old woman was referred for pelvic pain and irregular menses starting six months earlier. She had noticed abdominal distention three months before presentation.

On clinical examination, a large hypogastric mass was palpable. Ultrasonography showed a huge heterogeneous predominantly solid pelvic mass with some cystic foci. The patient was hospitalized with the diagnosis of a malignant ovarian tumor. All laboratory tests including tumor markers and serum hormonal assays were normal. The patient underwent bilateral salpingo-oophorectomy with no intraoperative pathologic diagnosis. Gross examination showed an encapsulated 23×8×8 cm mass weighing 2200 g attached to the right ovarian remnant and salpinx. On cut sections, the mass was solid with rubbery consistency and multiple degenerated cystic foci. The left salpingo-ovary was unremarkable.

Microscopic examination showed pseudolobulation, sclerosis, prominent vascularity, and two cell populations of spindle and polygonal cells (Figures 1–3).

Discussion

SST of the ovary is a rare tumor derived from the sex cord stroma. This tumor was first described by Chalvaridjian and Scully (1973), and occurs most frequently in the second and third decades of life. The tumor is usually hormonally inactive although some cases with irregular menses and genital bleeding have been reported. Peng et al.
found 114 cases reported until 2003.6

The patient reported here had no clinical virilization with normal serum hormone levels and normal blood tumor markers. However, a few cases with elevated serum CA-125 and hormonal abnormalities have been reported.1–3 It is difficult to differentiate SST from a malignant tumor in imaging studies.7 Nonetheless, some authors believe that preoperative diagnosis may be possible using magnetic resonance imaging which shows low-intensity nodules against a high-intensity stroma.6 Ultrasonographic findings in our patient were suggestive of malignancy and thus she underwent inappropriate surgery.

The histologic features include a pseudolobular pattern with focal areas of sclerosis and two distinct cell populations of spindled and polygonal cells.8 Also, fatty drops in the cytoplasm are positive with fat staining. Rarely, the lutein cells (polygonal cells) in SST evolve into a signet-ring-like9 structure that mimics the Krukenberg tumor which can be differentiated through immunohistochemical (IHC) tests and positive fat staining.

SST is positive for desmin and smooth muscle actin (SMA). The vascular endothelial growth factor is positive for both cellular and edematous areas. Other stromal tumors, i.e. thecoma and fibroma, tend to occur in the fifth and sixth decades,7 whereas almost 80% of SSTs occur in women under 30 years of age.9

The present case suggests that it is necessary to rule out SST in young patients and to confirm the diagnosis by intraoperative pathologic examination before selecting the treatment.

References